IDIOPATHIC INTRACRANIAL HYPERTENSION

PRESENTED BY NAA NAAMUAH TAGOE AT OSG FIRST CPD FOR 2017
GMA HOUSE
27-May-2017

GOALS

At the end of this talk, I expect all present

- To have a clear definition of IIH as well as knowledge of risk factors implicated in it.
- Provide a practical guide to evaluation and management of patients with idiopathic intracranial hypertension
- Understand that management of IIH is a multidisciplinary one and to comprehend the rational and indications for the various treatment

FORMAT

- Introduction to IHH
- Definition
- Epidemiology
- Risk factors
- Pathophysiology
- Work up of patients
- Treatment and follow up
- Prognosis
- Summary

INTRODUCTION

- Idiopathic intracranial hypertension(IIH) previously known as “Pseudotumor cerebri” or “benign intracranial hypertension” is a pathological state characterized by an increase in intracranial pressure with no obvious intracranial pathological processes
- It’s link with obesity and weight loss has led to an increase in the prevalence with a rise in worldwide obesity rates

Manfield JH et al Obes Surg. 2017 Feb;27(2):513-52

- Severe morbidity due to intractable, disabling headaches with significant risk of severe and permanent visual loss seen in up to 30%

Yri HM et al Neurol. 2011;259(5):886–94
Andrews LE, Liu GT, Ko MW. Idiopathic intracranial hypertension and obesity. Horm Res Paediatr. 2014;81(4):217–25.

DEFINITION

- Idiopathic intracranial hypertension (IIH) is a clinical syndrome defined by criteria that comprise symptoms and signs of intracranial pressure (e.g. headache, papilloedema and visual loss), elevated intracranial pressure (e.g. on lumbar puncture) with normal cerebrospinal fluid (CSF) composition and without any other cause identified on neuroimaging or other evaluations

Friedman DI et al. Neurology 2013; 81:1159–65.

- Not benign as previously thought since it causes devastating sequelae such as permanent visual loss in approximately 30-31% of cases, hence the need to make the diagnosis early and institute interventions

Andrews LE et al. Horm Res Paediatr. 2014;81(4):217–25.

Digre KB et al. Neurologist. 2001; 7:2–67.

DIAGNOSIS CRITERIA IIH

1. Required for the diagnosis of IIH

A. Papilledema
B. Normal neurologic examination except for cranial nerve abnormalities

C. Neuroimaging: **Normal brain parenchyma** without evidence of hydrocephalus, mass, or structural lesion and no abnormal meningeal enhancement on MRI, with and without gadolinium, for typical patients (female and obese), and **MRI,** with and without gadolinium, and magnetic resonance venography **(MRV)** for others.
If MRI is unavailable or contraindicated, contrast-enhanced CT may be used

D. Normal CSF composition

E. Elevated lumbar puncture opening pressure (≥250 mm CSF in adults and ≥280 mm CSF in children [250 mm CSF if the child is not sedated and not obese]) in a properly performed lumbar puncture

Friedman DI et al. Neurology 2013;81:1159–65.

Diagnosis of IIH without papilledema

In the **absence of papilledema**, a diagnosis of IIH syndrome can be made if B–E from above are satisfied, and in addition the patient has a unilateral or bilateral abducens nerve palsy

In the **absence of papilledema or sixth nerve palsy**, a diagnosis of IIH syndrome can be suggested but not made if B–E from above are satisfied, and in addition at least 3 of the following neuroimaging criteria are

i. Empty sella
ii. Flattening of the posterior aspect of the globe
iii. Distention of the perioptic subarachnoid space with or without a tortuous optic nerve

iv. Transverse venous sinus stenosis

A **diagnosis of IIH is definite if the patient fulfills criteria A–E.** The diagnosis is considered **probable** if criteria A–D are met but the measured CSF pressure is lower than specified for a definite diagnosis.

Friedman DI et al. Neurology 2013;81:1159–65.

EPIDEMIOLOGY

- Global incidence IIH variable from region to region according to incidence of obesity
- Increasing in prevalence in past decade, following obesity(BMI >30) epidemic

Arielle Spitze et al. Indian J Ophthalmol. 2014 Oct; 62(10): 1015–1021

- Estimated incidence of 1-3 people per 100,000 people per year, and occurs most commonly in obese, young women

Piper RJ et al. Cochrane Database Syst Rev. 2015 Aug 7;(8)

- Worldwide burden of idiopathic intracranial hypertension (IIH) continues to rise with current annual incidence estimated at up to 21 per 100,000 per year in obese young women

Manfield JH et al . Obes Surg. 2017 Feb;27(2):513-52

EPIDEMIOLOGY

- USA > 1/3 of adults are obese, compared with around 11% worldwide ; 1/3 overweight (body mass index (BMI) 25–30 kg/m^2)
- UK, 22.7% of people are obese

Mollan SP et al. Pract Neurol 2014;0:1–11.

- Prevalence of IIH in USA
  - 0.9-1.0 per 100 000 in general population
  - 1.6-3.5 per 100 000 in women
  - 7.9- 20 per 100 000 in overweight women

Eye wiki 2015 feb

WHAT'S THE PICTURE IN AFRICA/GHANA

- Prevalence of overweight/obesity(adults) Ghana -43%
  - overweight 25.4% (95% CI 22.2–28.7%)
  - obesity 17.1% (95% CI = 14.7–19.5%)
  - women > men (p<0.0001)
- Regional level, about 43.4%, 36.9%, 32.4% and 55.2% residents in Ashanti, Central, Northern and Greater Accra region, respectively are overweight or obese.

Richard Ofori-Asenso et al. BMC Public Health 2016;16:1239: 2-18

IIH IN AFRICA AND GHANA

Case reports in Nigeria and South Africa

KBTH eye department

2008 - 2014 – 4 cases

2015/ Sept 2016 – 3 cases

Oct 2016- date – 6 confirmed,2 probable

SUMMARY OF WEIGHT/BMI

CASE	Age	Gender	WEIGHT(KG)	HEIGHT(m)	BMI
1	31	female	Not done	Not done	
2	8	female	46	1.3	27.2 Overweight
3	20	female	124	Not done	
4	21	female	Not done	Not done	
5	24	female	100	1.73	33.4 Obese
6	40	female	97	1.53	40.9 Obese

DURATION OF SYMPTOMS/VA

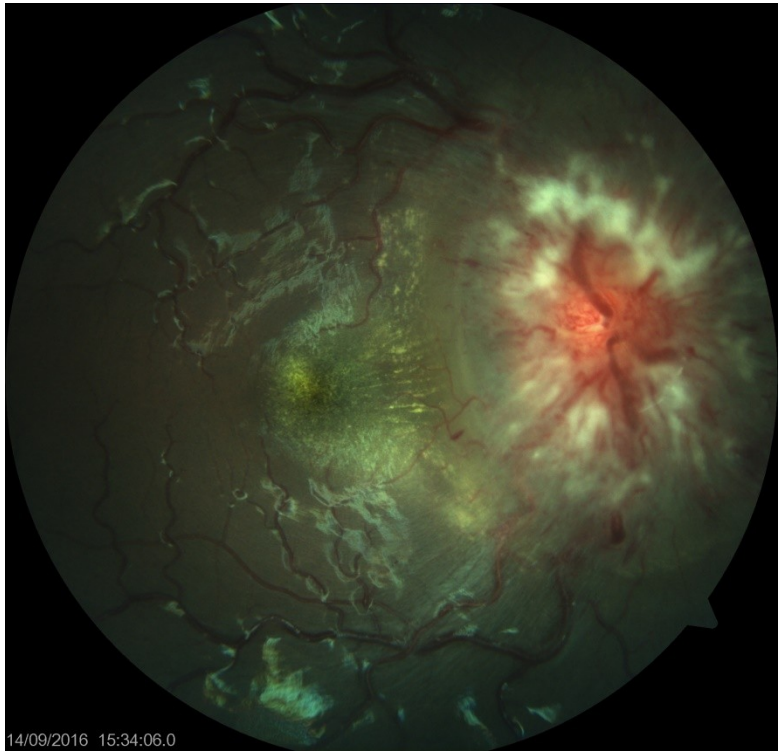
CASE	DURATION OF SYMPTOMS	INITIAL VISUAL ACUITY		VA AFTER I/12 OF RX		VA AFTER 3/12	
		RE	LE	RE	LE		
1	2/52	6/9	6/9	6/9	6/9	6/6	6/6
2	4/12	6/36	6/36	6/24	6/9	6/9	6/6
3	1/12	6/36	6/12	6/18	6/12	X	X
4	UNSPECIFIED	6/6-1	6/6-1	X	X	X	X
5	3/52	6/6	6/9	X	X	X	X
6	3/12	6/6	6/6	6/6	6/6	6/6	6/6

Lost to follow up

CASE	BLOOD PRESSURE /mmHg	GRADE OF PAPILLOEDMA (FRISEN)	CSF OPENING PRESSURE	OPTIMUM DOSE OF ACETAZOLAMIDE (DOSE AT WHICH SYMPTOMS RELIEVED)
1	140/80	4	NOT DONE	250MG BD
2	100/60	5	Done but not measured!	500MG TID
3	149/96	5	35CMH2O	500MG QID
4	NOT DONE	3	55CMH2O	500MG BD
5	120/72	3	NOT DONE	500MG TID
6	138/85	3	NOT DONE	500MG TID

CASE 2 (RIGHT EYE)

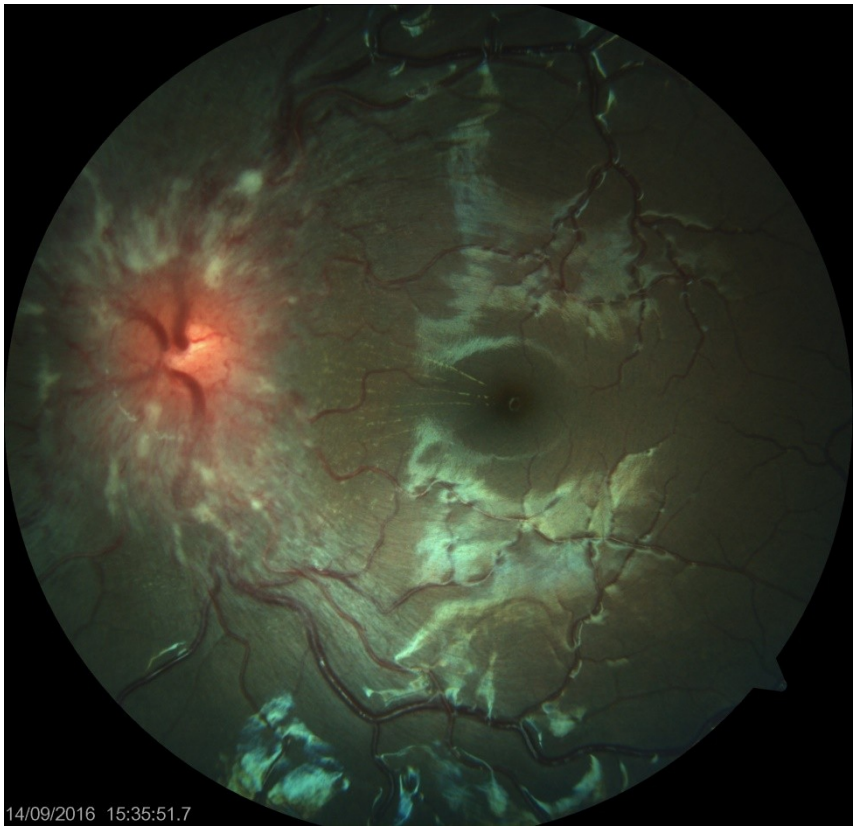
At presentation



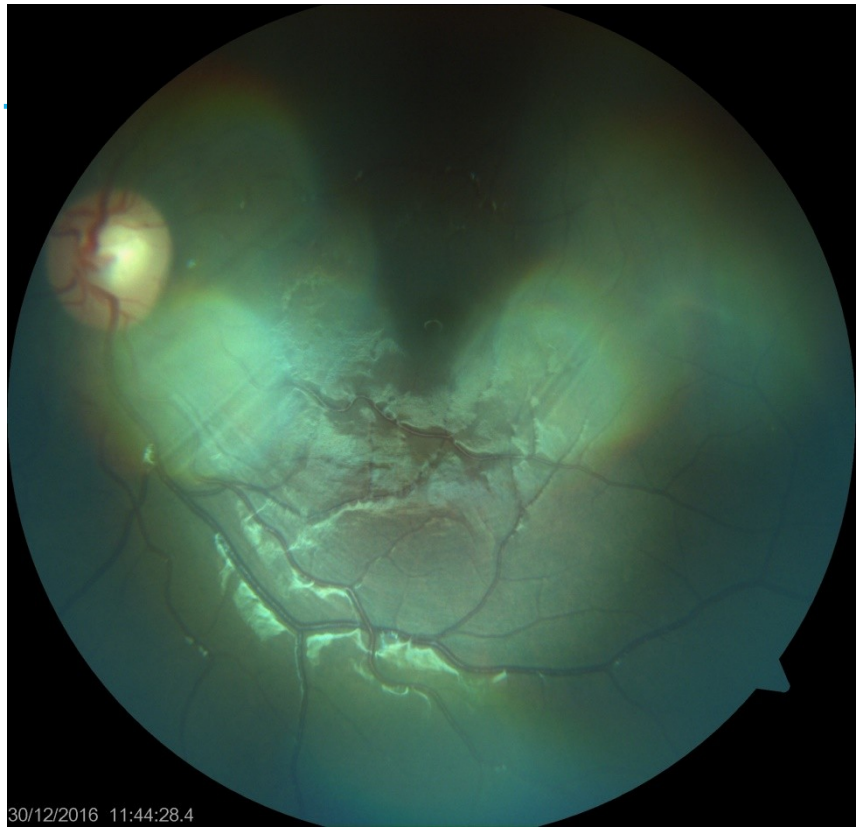
3 months post treatment

CASE 2 (LEFT EYE)

At presentation



3 months post treatment



RISK/PRECIPITATING FACTORS

Systemic illnesses

- Obstructive sleep apnea
- Hypothyroidism
- Anaemia
- Addison’s disease
- Systemic lupus erythematosus
- Behcet’s syndrome
- Polycystic ovarian syndrome
- Coagulopathies
- Uraemia

Medications

- Oral contraceptive pills
- Vitamin A
- Tetracycline
- Nalidixic acid
- Cyclosporin

CLASSIFICATION

- As a result of precipitating factors listed terms "**primary**" and "**secondary**" intracranial hypertension suggested to describe

  - young obese women with isolated raised ICP and no obvious precipitating factors
  - patients with isolated raised ICP associated with factors such as endocrine disorders, anemia, obstructive sleep apnea, medications, or cerebral venous sinus stenoses.

Friedman DI et al. J Neuroophthalmol. 2004; 24:158-145.
Digre KB. Neuroophthalmology.2009; 33:93–99.

PATHOPHYSIOLOGY-THEORIES

- Remains idiopathic but is most likely due to ***increased CSF production, reduced CSF absorption, increased cerebral venous pressure, venous sinus stenosis, increased brain water content***, or a combination of these
- Cerebral edema – evidence against is no focal neurological signs or altered level of consciousness, alertness
- Increase intrabdominal pressure secondary to obesity- Impedes venous return from brain.
- Non-obese patients (BMI<30) at >risk with recent weight gain of 5-15% of body weight.

Biousse V. et al. J Neurol Neurosurg Psychiatry. 2012;83:488–94.

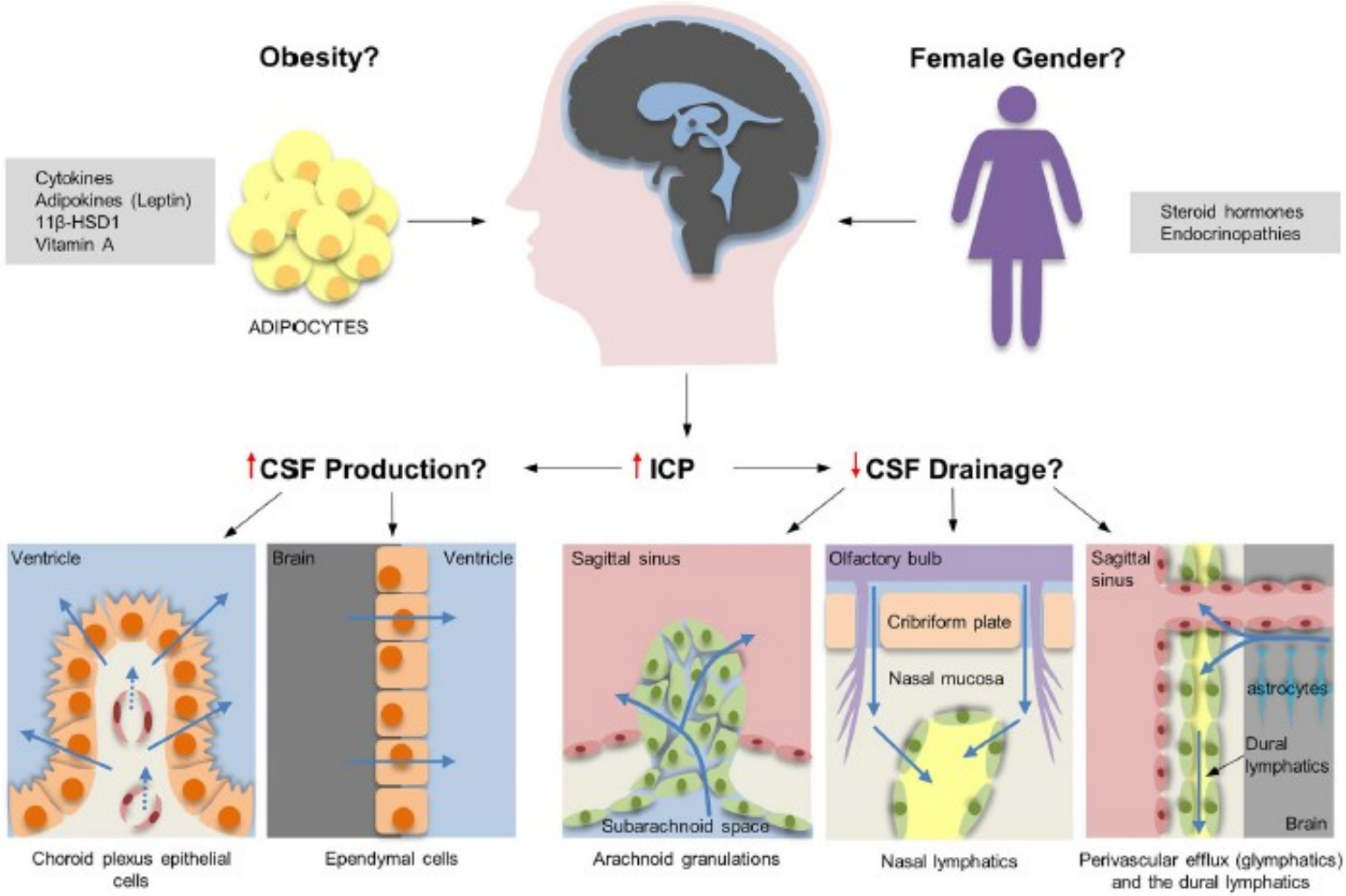
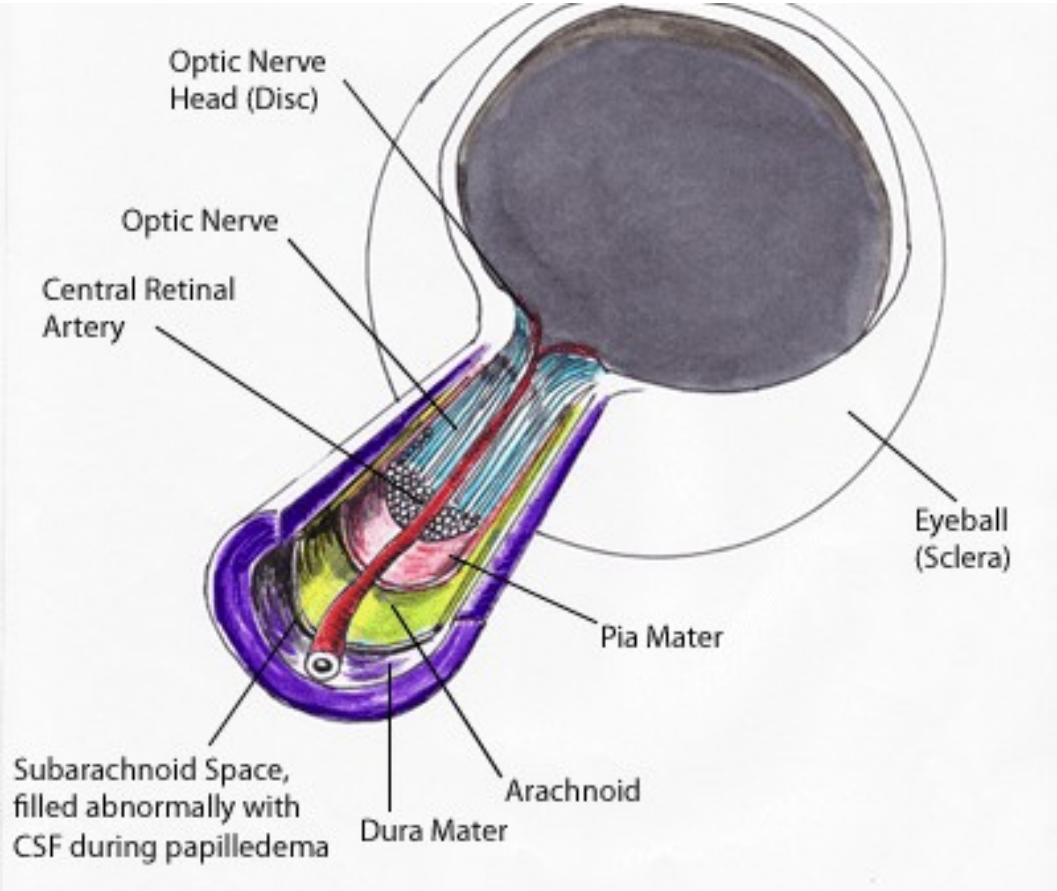
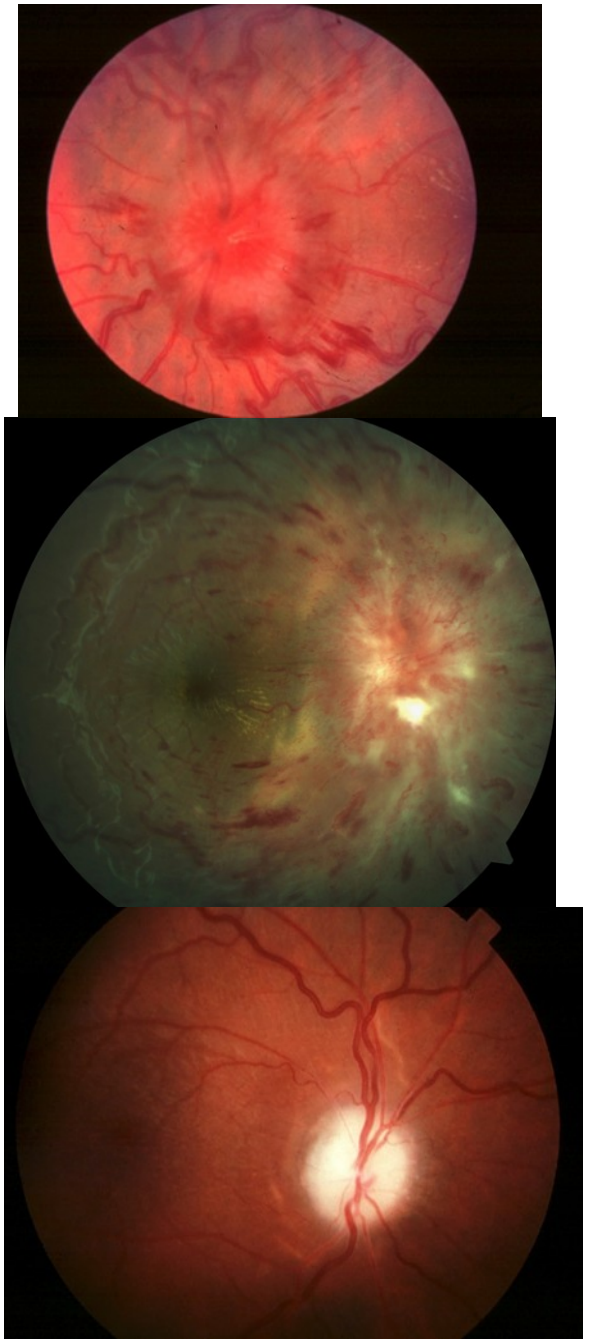


Figure 1 Schematic diagram of the possible pathophysiological mechanisms in idiopathic intracranial hypertension (IIH). Cerebrospinal fluid (CSF) is produced mainly by the choroid plexus epithelial cells, with a small amount being secreted by ependymal cells that line the ventricular system. Classically, CSF was thought to drain predominantly through the subarachnoid space through arachnoid granulations into the superior sagittal sinus. Evidence also suggests CSF drains through the cribriform plate along cranial nerves into the nasal lymphatics (yellow). The most recent hypothesis proposes bulk flow of fluid along perivascular routes (glymphatic pathway) which is cleared from the brain into the subarachnoid CSF, bloodstream or cervical lymphatics. Supporting this concept is the recent discovery of lymphatic vessels (yellow) in the dura that drain into the deep cervical lymph nodes.

 Moolan SP et al . J Neurol Neurosurg Psychiatry 2016;87:982–992

OPTIC NERVE


Optic Nerve Head (Disc)
Optic Nerve
Central Retinal Artery
Eyeball (Sclera)
Pia Mater
Subarachnoid Space, filled abnormally with CSF during papilledema
Arachnoid
Dura Mater



WORK UP

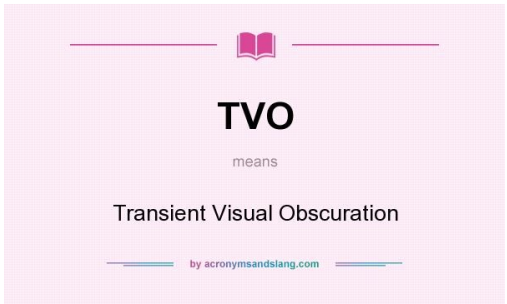
- History- symptoms, recent weight gain, drugs etc
- Complete ocular examination including VA, colour vision, a dilated fundus exam, cranial nerves
- General and neurological examination; check BP to rule out malignant hypertension
- Visual field
- Fundus photographs
- Neuroimaging
  - 1. MRI of head &orbit with contrast /MRV imaging of choice. Fat suppression images better define intraorbital optic nerve.
  - 2. CT& CT venogram in absence of MRI to exclude SOL and chiari malformations

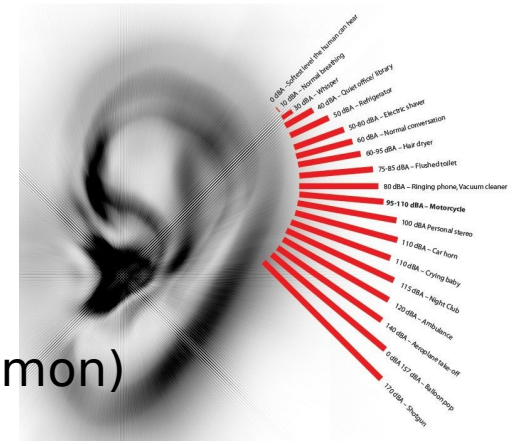
    to identify any space-occupying lesion

    exclude a venous sinus thrombosis
- Lumbar puncture – mandatory.

HISTORY

Headache





adicular pain in arm(uncommon)

Pulsatile tinnitus

HISTORY

Box 2 Secondary causes of raised intracranial pressure for exclusion to diagnose IIH

- Secondary causes of raised intracranial pressure.
- Venous sinus thrombosis.
- Anaemia.
- Obstructive sleep apnoea.
- Drug-related.
- CSF hyperproteinaemia/hypercellularity, for example, spinal cord tumour/meningitis/Guillain–Barré syndrome/subarachnoid haemorrhage.
- Renal failure.
- Endocrine diseases, for example, Addison's/Cushing's/hypothyroidism.

Mollan SP et al. Pract Neurol 2014;0:1–11

EXAMINATION

Normal level of consciousness but patient complains of severe headache

Visual loss(typically field but rarely acuity) except macular involvement or atrophic optic nerve

STAGING OF PAPILLOEDEMA

- Modified Frisen- 0-5
- Clinical – early, established, chronic , atrophic

STAGING OF PAPILLEDEMA

Early papilledema

VA normal

Disc hyperaemia, indistinct nasal margin, mild venous engorgement,

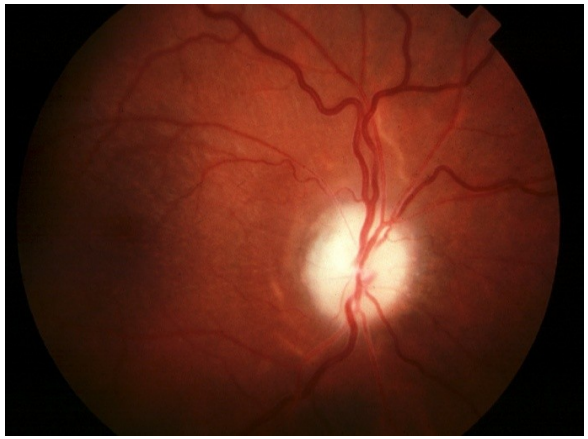
Normal cup, absent spontaneous venous pulsation.

VA normal, severe hyperaemia, indistinct margins, obscuration of small disc vessels,marked venous engorgement, absent cup, haemorrages/cotton wool spots, macular star

STAGING OF PAPILLEDEMA

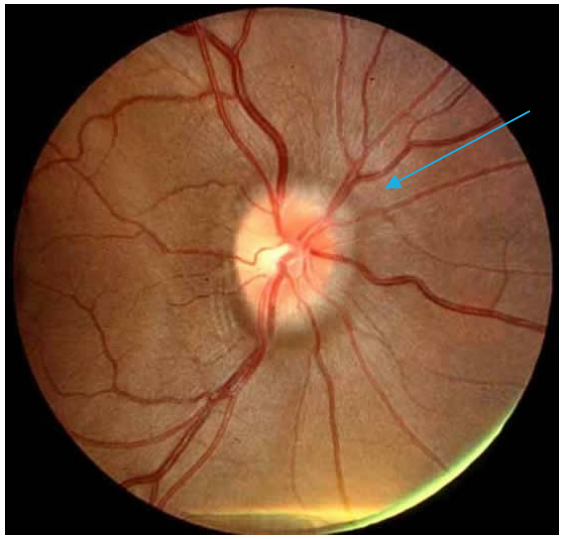
Long standing/chronic

VA variable , marked disc elevation, less hyperaemia, indistinct disc margin, variable venous engorgement, absent cup

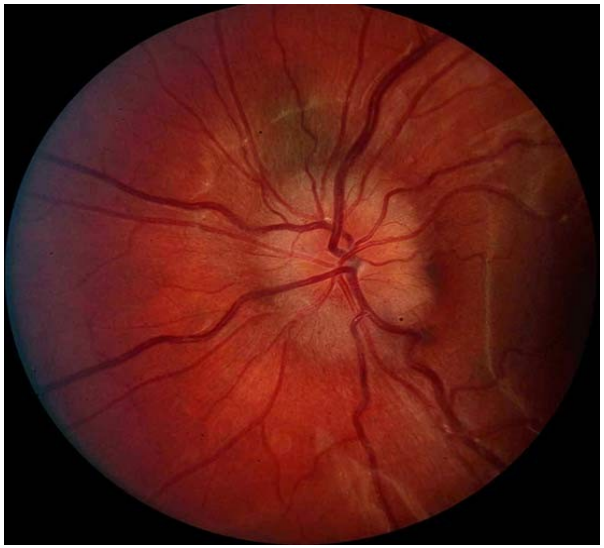


VA reduced, mild disc elevation , indistinct margins, disc pallor, few sclerosed vessels, absent cup

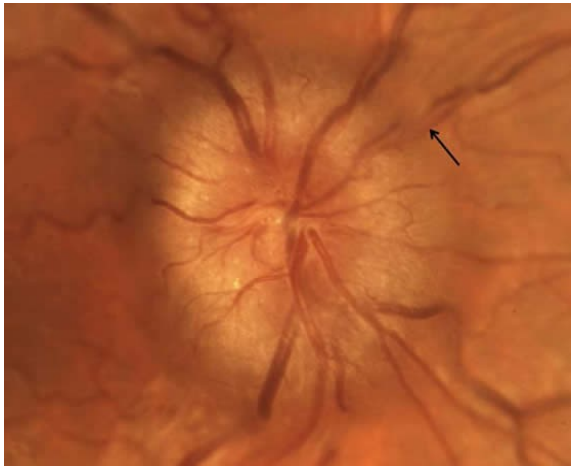
Grade 0



Grade 1 C-shaped halo



Grade 2 Circumferential ha



Grade 3
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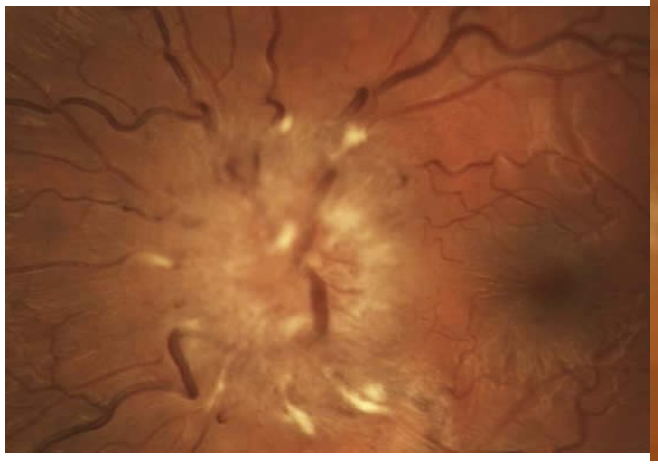
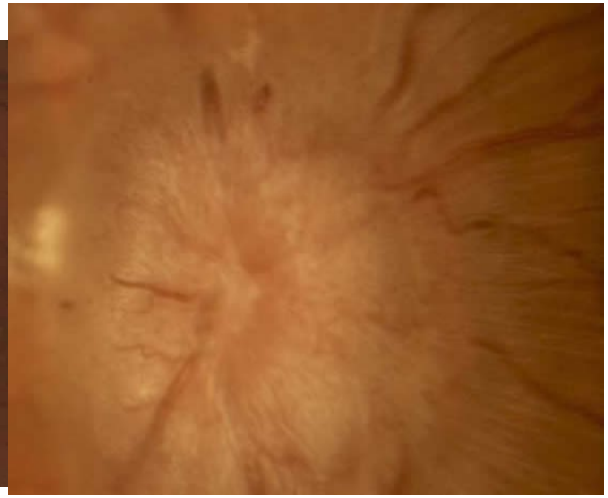
ve disc



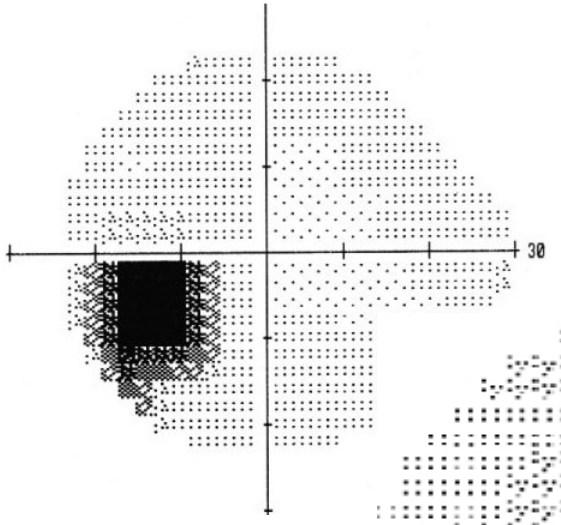
Grade 4 loss of major vessels on disc

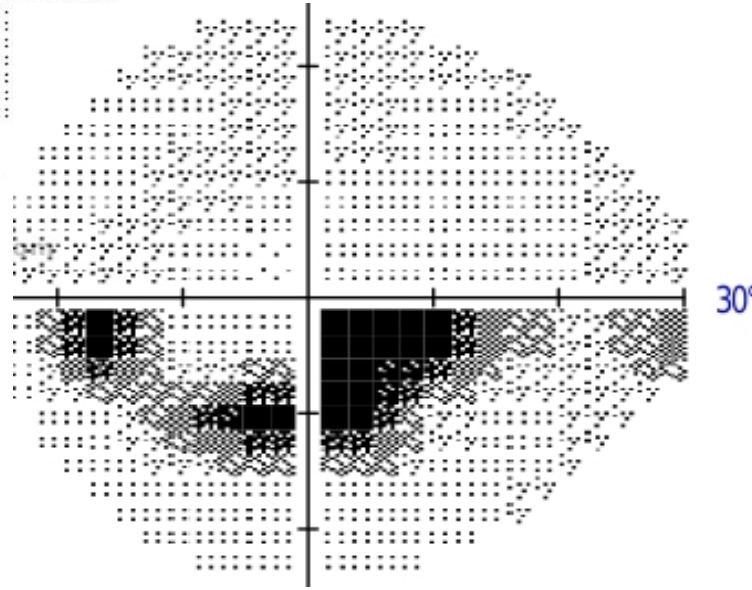


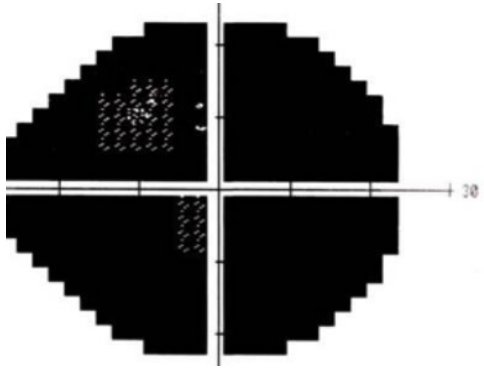
Grade 5, loss of all vessels

VISUAL FIELDS

Most common type of IIH baseline hemifield abnormality localized nerve fiber bundle-like defect. Localized inferior hemifield loss common than superior hemifield loss





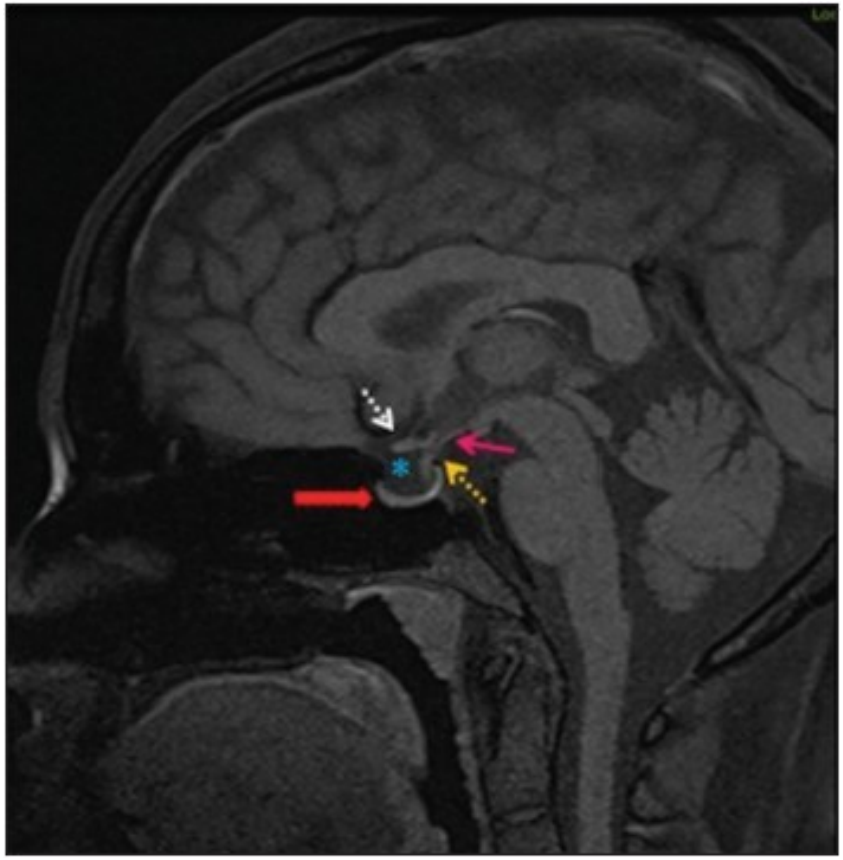


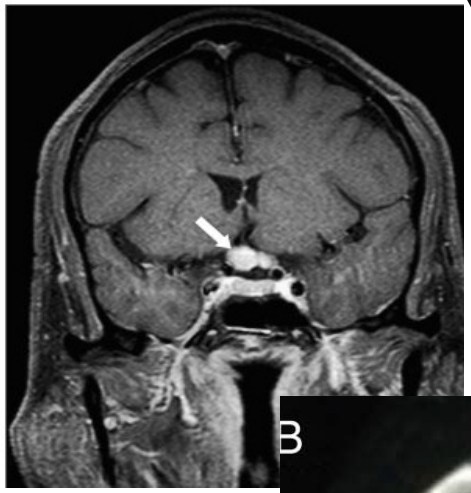
John L. Keltner, et al Invest Ophthalmol Vis Sci. 2014 May; 55(5): 3200–3207

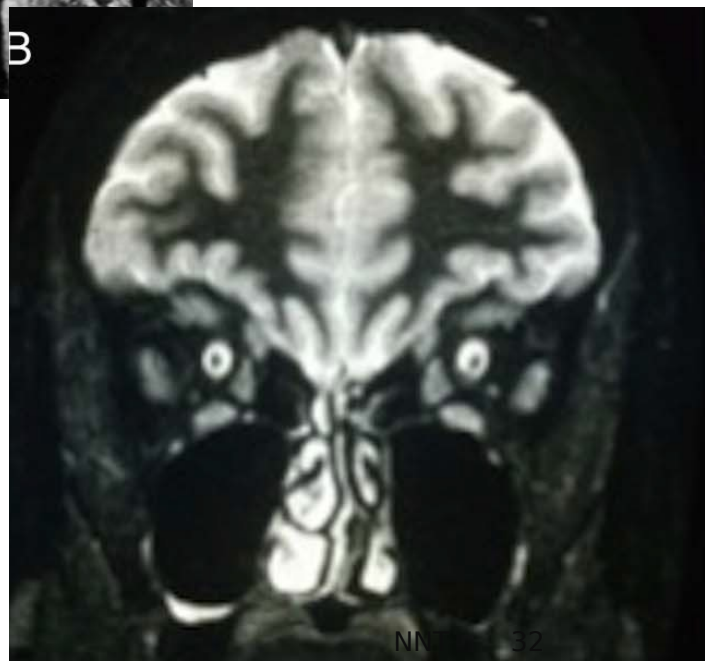
MRI FINDINGS

- Flattening of the posterior pole
- Distension of perioptic subarachnoid space
- Enhancement (with gadolinium) of the prelaminar optic nerve
- Empty sella
- Intraocular protrusion of the prelaminar optic nerve
- Vertical tortuosity of the orbital optic nerve
- Stenosis of one or both transverse cerebral venous sinuses

EMPTY SELLA
ENHANCEMENT OF NERVE





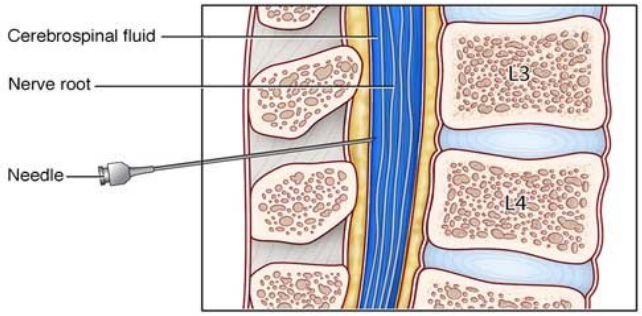


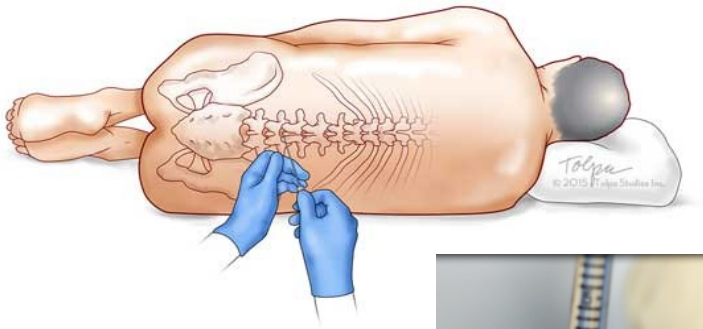
MRI FINDINGS

Flattening of posterior sclera

Vertical tortuosity of optic nerve

LUMBAR PUNCTURE





- Diagnosis based on opening pressure of > 250 mm of water in adult with patient lateral decubitus position .
- 200mm of water to 250 mm water equivocal
- CSF: bacteriology, biochemistry and cytology
- Patients with IIH should have normal or low protein

DIFFERENTIAL DIAGNOSIS

Any condition which would cause papilloedema ± headache

- Intracranial mass(tumour, abcess, haemorrhage)
- Increased CSF production eg choroid plexus tumour
- Decreased CSF absorption eg arachnoid granulation adhesions following infections, meningitis, subarachnoid haemorrhage
- Obstructive hydrocephalus
- Obstruction of venous outflow eg ***venous sinus thrombosis***, jugular vein compression

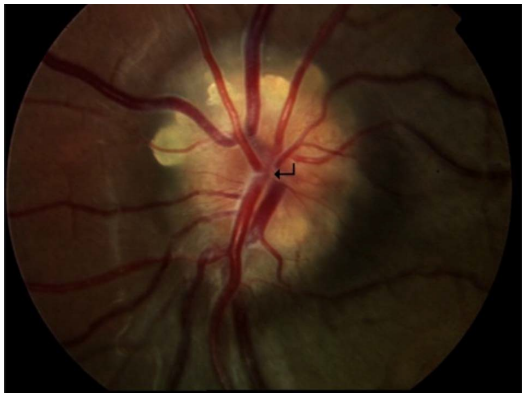
DIFFERENTIAL DIAGNOSIS

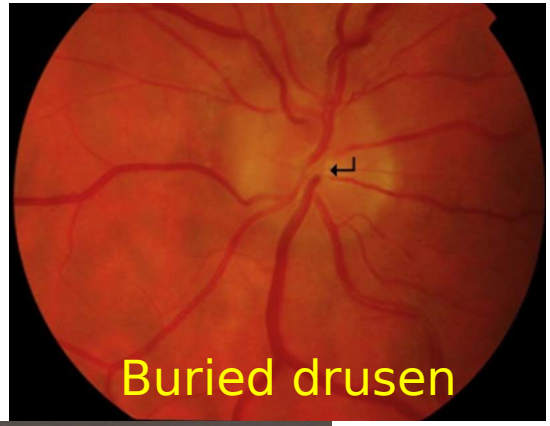
Diseases that would cause unilateral or bilateral disc edema

- Papillitis
- Hypertensive optic neuropathy
- Central retinal vein occlusion
- Ischaemic optic neuropathy
- Infilteration of optic disc
- Orbital optic nerve tumours
- Thyroid related optic neuropathy

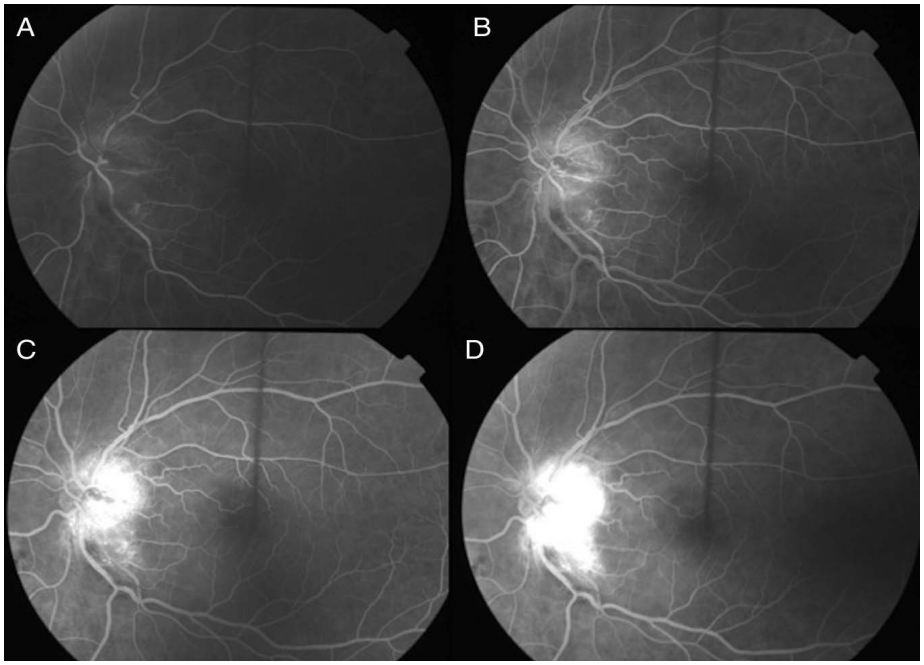
Pseudopapilledema – due to congenitally anomalous discs, optic

nerve head drusen or a combination

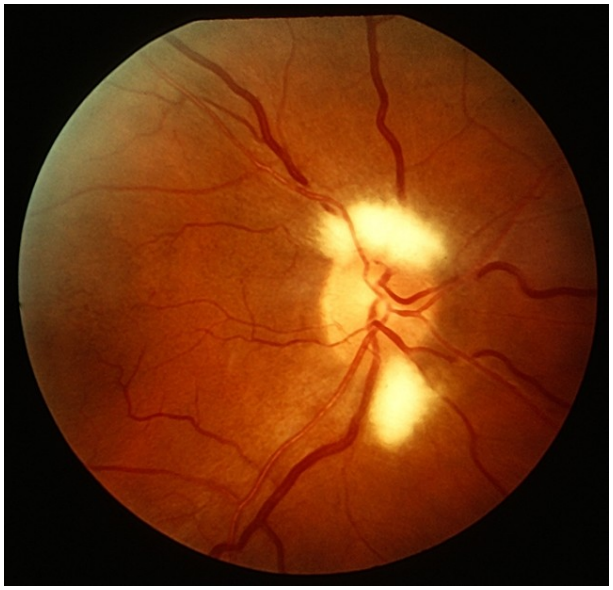


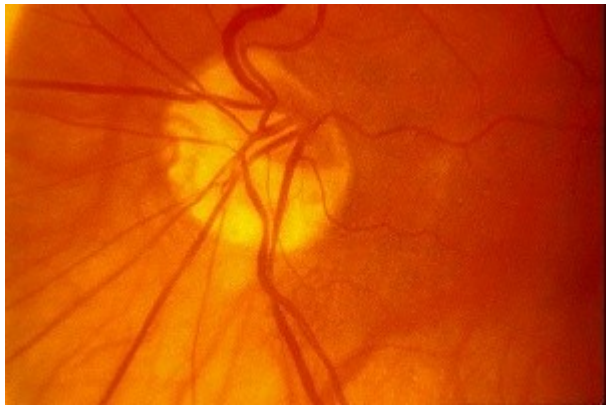


B-scan ultrasound of the right optic nerve, showing optic nerve head drusen. Ovoid echogenic foci in optic nerve

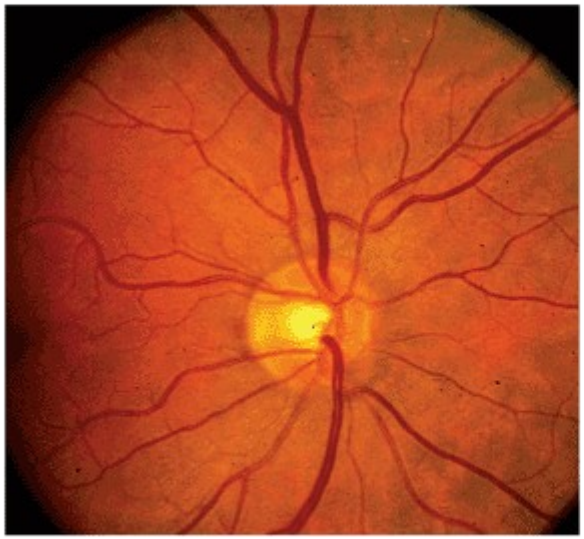


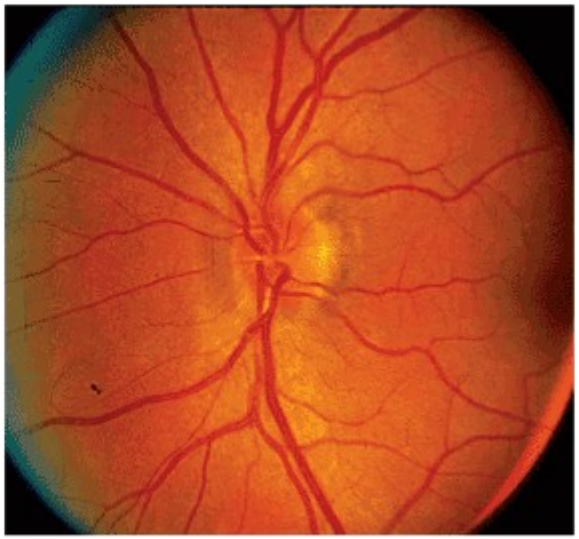
Leakage in true disc swelling





Tilted disc





Left: A normal disc; note the central cup. Right: A "disc at risk." Note that the disc is small, and there is no central cup.

GOAL OF TREATMENT

- **To alleviate symptoms of raised intracranial pressure and preserve vision.**

TREATMENT

- Multidisciplinary approach involving ophthalmologist, neurologist, neurosurgeon, radiologist, nutritionist

WEIGHT LOSS

- Only proven disease-modifying treatment for all overweight patients

Sinclair A, Burdon M, Ball A, et al. BMJ 2010;7:341

Newborg B. Arch Intern Med 1974;133:802–7

- 5-10% found to improve symptoms and signs
- Final absolute BMI is more important for patients stopping treatment rather than average weight loss.
- Bariatric surgery option in morbidly obese patients.

Wong R. et al. BMC Ophthalmology 2007, 7:15

WEIGHT LOSS

- 15% reduction of body weight, using a low calorie meal replacement liquid diet for 3 months

significantly reduced intracranial pressure, papilloedema and headaches.

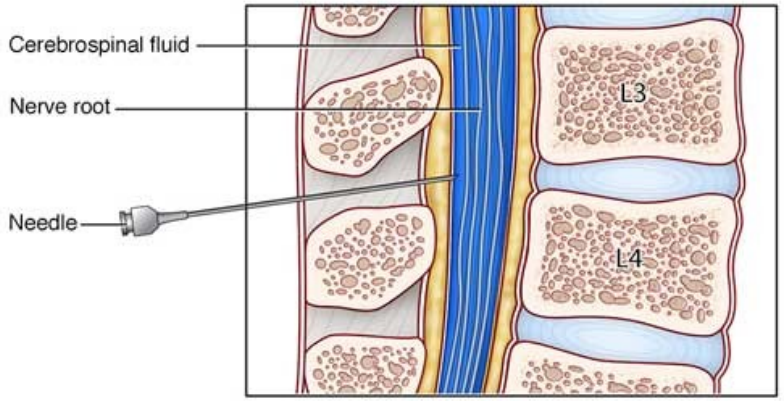
12. Sinclair A, Burdon M, Ball A, et al. BMJ 2010;7:341

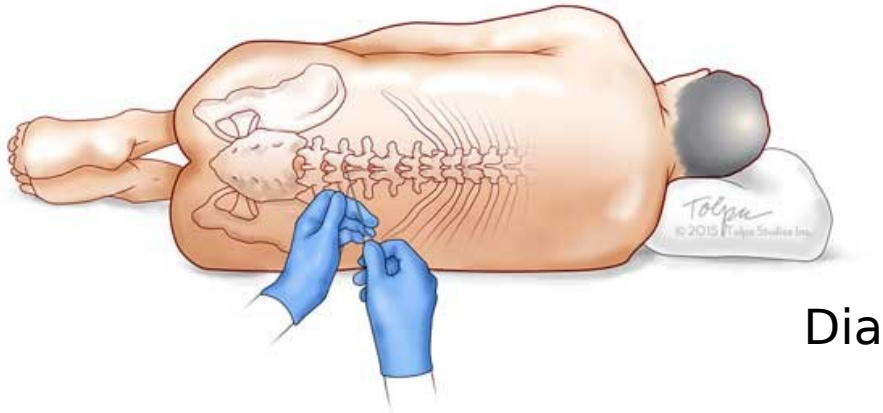
EVIDENCE FOR BARIATRIC SURGERY

- Option in morbidly obese with severe symptoms
- Achieved 100% papilledema resolution and a reduction in headache symptoms in 90.2%.
- Non-surgical methods offered improvement in papilledema in 66.7%, visual field defects in 75.4% and headache symptoms in 23.2%.
- Surgical BMI decrease was 17.5 vs. 4.2 for non-surgical methods

.**Manfield JH et al Bariatric Surgery or Non-surgical Weight Loss for Idiopathic Intracranial Hypertension? A Systematic Review and Comparison of Meta-analyses. Obes Surg.** 2017 Feb;27(2):513-52

LUMBAR PUNCTURE





Diagnostic/therapeutic

MEDICAL THERAPY

More beneficial in mild to moderate disease

CAUTION

- Currently no place for steroids due to long term undesired side effects (weight gain)

- Rebound intracranial hypertension ff withdrawal

Biousse V et al .J Neurol Neurosurg Psychiatry. 2012 May ; 83(5): 488–494.

ACETAZOLAMIDE-CARBONIC ANHYDRASE INHIBITOR

Child

- 25 mg/kg/day
- increased by 25mg/kg/day until clinical response or a maximum dose of 100 mg/kg/day or 2 g/day

Soler D. et al. Arch Dis Child. 1998 Jan;78(1):89-94

Adult

- 4 tabs (250 mg/tab) daily in 2 divided doses
- Increase by 250 mg weekly till

4 g dose attained

- acceptable safety profile at dosages up to 4 g/d in IIH.
- Majority of participants were able to tolerate acetazolamide above 1 g/d for 6 months

Ten Hove MW et al J Neuroophthalmol. 2016 Mar;36(1):13-9

- Dose of 1g/d adequate ; modified release preparation helps reduce side effects.

Mollan SP et al. Pract Neurol 2014;0:1–11

A CASE FOR MEDICAL THERAPY VRS WEIGHT REDUCTION –NORDIC IIHTT

- Aim: determine beneficial effect of acetazolamide in improving vision when added to low-sodium weight reduction diet in patients with IIH and mild visual loss
- **Multicenter, randomized, double-masked, placebo-controlled** study
- **165 participants from 38 academic centres** with IIH and mild visual loss who received a low-sodium weight-reduction diet.
- North America :March 2010 to November 2012
- Follow up for 6 months
- All participants met the modified Dandy criteria for IIH and had a perimetric mean deviation (PMD) between −2 dB and −7 dB.
- Mean age 29 years and all but 4 participants were women.

NORDIC IIH study group JAMA April 23/30, 2014 :311(16): 1641-1651

RESULTS

- Intervention : Low-sodium weight-reduction diet plus maximally tolerated dosage of acetazolamide (up to 4 g/d) or matching placebo for 6 months
- Primary outcome variable: change in PMD from baseline to month 6 in most affected eye, by Humphrey Field Analyzer.
- Conclusion : **Use of acetazolamide with a low-sodium weight-reduction diet compared with diet alone resulted in modest improvement in visual field function in IIH patients with mild visual loss**

NORDIC IIH study group JAMA April 23/30, 2014 :311(16): 1641-1651

- Marked reductions in baseline QOL seen among patients with mild visual loss from IIH are improved by treatment with acetazolamide

.Bruce BB._ Neurology. 2016 Nov 1;87(18):1871-1877. Epub 2016 Sep 30.

EVIDENCE FOR TOPIRAMATE

- 40 patients single centre with IIH
- Randomly assigned to acetazolamide (1-1.5g/d) or topiramate(100 or 150mg/d)
- prospectively open label study.
- Demographic characteristics, clinical features CSF pressure similar at beginning of study.
- Primary endpoint change visual field defect grades at 3, 6 and 12 months.

Celebisoy N et al Acta Neurol Scand 2007: 116: 322-327.

EVIDENCE FOR TOPIRAMATE

- Secondary end points papilledema, transient visual obscurations, diplopia and headache. Assessment of outcomes was not blinded.
- Results - topiramate was as effective as acetazolamide in relieving headache.
- Patients taking topiramate lost statistically significantly more weight than patients taking acetazolamide
- Conclusion : Topiramate effective in treatment of IIH.
- Weight reduction as well as the reduction of the CSF formation is possible mechanism of action.

Celebisoy N et al. Acta Neurol Scand 2007: 116: 322-327.

SURGERY INDICATIONS

- Development of new visual field defect
- Worsening of previous visual field defect
- Severe visual loss at time of presentation
- Psychosocial ie non adherence to medication

Corbett JJ et al Arch Neurol 1989:46(10): 1049

- Refractory headaches.

SURGICAL OPTIONS AVAILABLE

- Optic nerve sheath fenestration

  (ONSF)

- CSF Shunting procedures
  - Lumbo peritoneal shunt
  - Ventriculo-peritoneal shunt

CSF SHUNTING PROCEDURES

- Not recommended exclusively to treat headache, since these continue in most patients postoperatively (68% at 6 months and 79% at 2 years)
- Postoperative low-pressure headache occurs in 28%.

Sinclair AJ et al Cephalagia 2011;31:1627-33

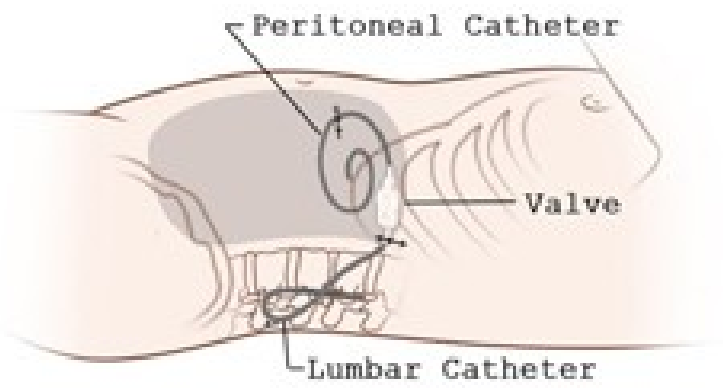
- Main challenge is malfunction, over-drainage, infection.
  - Shunts with a valve system and CSF reservoir to reduce the morbidity from over-drainage and under-drainage

Mollan SP, et al. Pract Neurol 2014;0:1–11

- Half of patients need shunt revision and a third multiple revision

Sinclair AJ et al. Cephalagia 2011;31:1627-33

LUMBO PERITONEAL SHUNT (LPS)



COMPLICATIONS

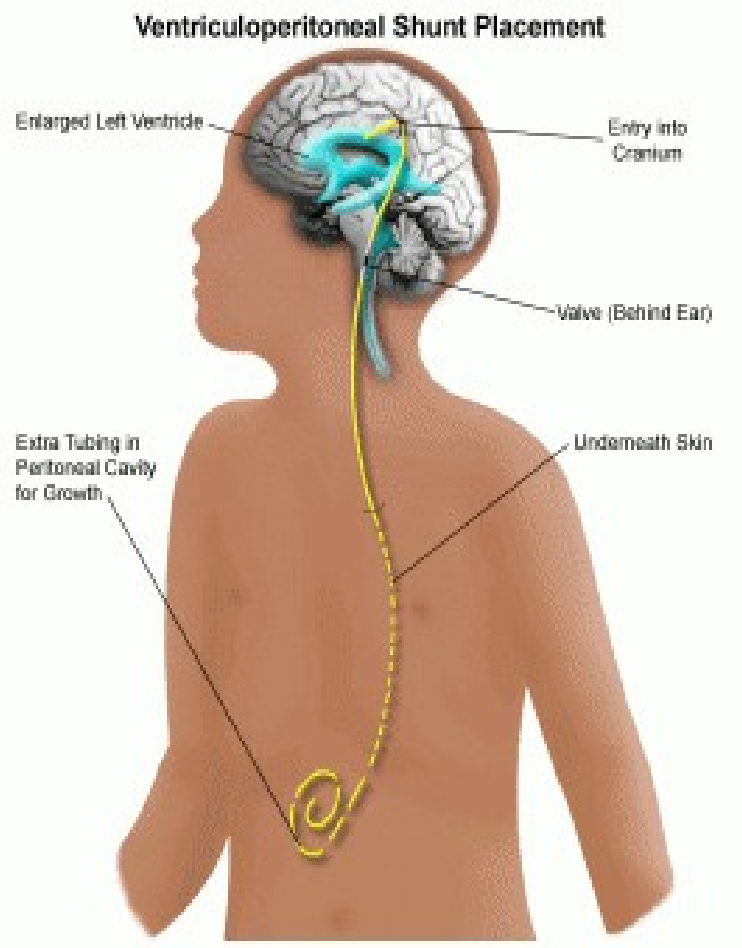
- Shunt malfunction
- Low pressure headaches
- Acquired cerebellar tonsillar
- Herniation
- Lumbar radiculopathy
- Infection

VENTRICULO PERITONEAL SHUNT(VPS)

- Not widely used for IIH because of difficulty placing shunt into normal or small-sized ventricles.
- Effectiveness and safety greatly increased with advent of image guidance.

COMPLICATIONS

- Shunt obstruction
- Low pressure headaches
- acquired cerebellar tonsillar
- Herniation
- Infection



VP VERSUS LP SHUNTS

- Revision rates were higher with LP shunts (60%) than with VP shunts (30%)

. ***Biousse V et al J Neurol Neurosurg Psychiatry. 2012 May ; 83(5): 488–494***.

EVIDENCE FOR VP SHUNT OVER LPS

- 18% to 85% complication rate with lumboperitoneal (LP) shunts
  - multiple revision surgeries
  - iatrogenic Chiari malformation
  - frequent wound complications

- 4480 patients with IIH identified
  - 2505 undergoing first-time VP shunt placement
  - 1754 undergoing initial LP shunt placement.

Menger R P et al Neurosurg Focus 2014; 37(5):

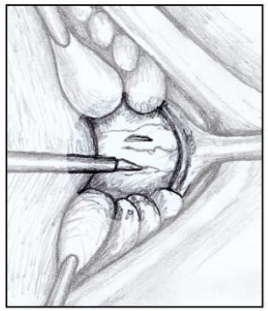
EVIDENCE FOR VP SHUNT OVER LPS

- Revision surgery occurred in
  - 3.9% of admissions (n = 98) for VP shunts
  - 7.0% of admissions (n = 123) for LP shunts ($p < 0.0001$).

- At teaching institutions preferred first time shunts for IIH were
  - VP shunts 83.8% of cases
  - LP shunts 77.3% of cases. ($p < 0.0001$)

24 Menger R P et al A comparison of lumboperitoneal and ventriculoperitoneal shunting for idiopathic intracranial hypertension: an analysis of economic impact and complications using the Nationwide Inpatient Sample Neurosurg Focus 2014; 37(5):

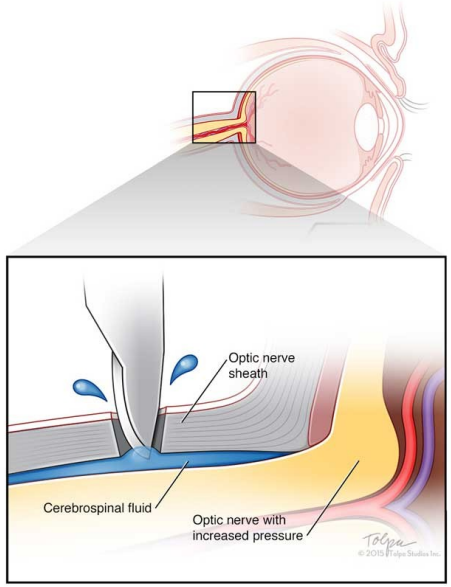
OPTIC NERVE SHEATH FENESTRATION(ONSF)



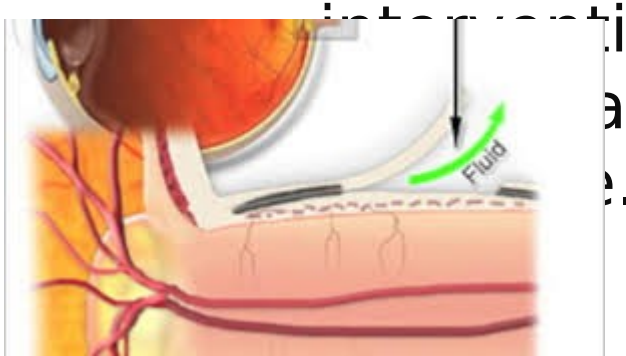
Incision in dura
Covering optic nerve

- Indication-
  - severe papilloedema with visual loss but no ICP symptoms
  - Patients with renal failure
  - Visual loss during pregnancy

Soler D. et al. Arch Dis Child. 1998 Jan;78(1):89-94



- Reverses or preserves vision in 80-90%
- key to success-early int ion/ ate e.



OPTIC NERVE SHEATH FENESTRATION(ONSF)

- Role in management of IIH controversial

- Indications, risks, and benefits compared to CSF diversion procedures not fully elucidated.
- Expertise of surgeon important

- Retrospective record review of 37 patients (50 eyes) had ONSF by a single surgeon

Pinneles S. et al. Neuro-ophthalmology 2013;37(1):12-19

OPTIC NERVE SHEATH FENESTRATION

- Visual acuity (VA) improved in 22% of operated eyes and 17% of fellow eyes
- stabilized in 54% of operated and 74% of fellow eyes
- deteriorated in 24% of operated and 9% of fellow eyes.
- Better pre-operative VA ($p = 0.01$), colour vision ($p = 0.002$), and earlier intervention ($p = 0.04$) associated with stabilization.
- Conclusion- *ONSF often stabilizes vision and visual fields. Results best in patients with better pre-operative vision and in those with earlier intervention*

Pinneles S. et al. Neuro-ophthalmology 2013;37(1):12-19

OPTIC NERVE SHEATH FENESTRATION

- Banta and Farris described a 94% improvement in visual acuity (148 of 158 eyes)
- 88% rate of visual field stabilization or improvement (71 of 81 eyes) after ONSF. There was only one eye (<1%) that suffered any severe, vision-limiting surgical complications.

Banta JT et al. Ophthalmology. 2000 Oct; 107(10):1907-12

MANAGING HEADACHE

- Characteristics very variable, may not fit classic described by International Headache Society

Moolan et al. Pract Neurol 2014;0:1–11

- Headaches usually multifocal
  - raised intracranial pressure
  - low pressure
  - medication overuse headache
  - migraine or a combination of these.
- Chronic disease – severe headache despite resolved papilloedema

D'Amico D, et al. Neurol Sci 2013;34:147–49.
Friedman DI et al. Neurology. 2002;58:1551–3.
Gonzalez-Hernandez A et al. Rev Neurol 2009;49:17–20.
D'Amico D, et al. Neurol Sci 2012;33:189–91.

MANAGING THE HEADACHE

- Migrainous features common (>70% of patients)

D'Amico D, et al. Neurol Sci 2012;33:189–91.

- Evaluate and treat headache type.
- Headache diary helpful for both patient and clinician
- Topiramate very useful . Occasionally amityptilline

Mollan SP, et al. Pract Neurol 2014;0:1–11.

IIH IN CHILDREN

- Obesity not risk factor in prepubertal cases
- Most cases are secondary IIH

Abdulrahman A. Journal of Paediatrics 2016; 16(2):67-76

- Similar symptoms but in addition lethargy and tiredness, dizziness, mood change, and intracranial buzzing sounds.
- Sleep and behaviour disturbances are often reported by parents in the young preverbal child. level of consciousness and intellectual functioning remains normal

Soler D. et al. Arch Dis Child. 1998 Jan;78(1):89-94

- Sixth nerve palsy is the most common neurological abnormality reported in 9–48% of children with BIH

Abdulrahman A. Journal of Paediatrics 2016; 16(2):67-76

- 280 mmH2O is considered as the upper limit of CSF opening pressure in children between 1 and 18 years

Abdulrahman A. Journal of Paediatrics 2016; 16(2):67-76
Lee MW et al .Pediatr Neurol. 2011;45(4):238-40.

IIH IN CHILDREN

- Treatment is indicated when there is evidence of **visual loss, moderate to severe papilledema, or persistent headaches**
- Mild asymptomatic papilledema- observe
- Goals of treatment and options similar to adults
- Duration of treatment variable but may be as long as 14 months

MONITORING IIH

Ocular

- Symptoms of patient- new/improved/resolved.
- Visual acuity
- Colour vision
- Serial visual fields
- Colour fundus photos- disc/macula
- OCT

Non ocular

- Height
- Weight
- BMI
- BP

COURSE OF DISEASE

- Rapid loss of vision at diagnosis over days to weeks (rare but vital to identify early).
- Disease resolution following diagnosis, over weeks to months, occasionally after a single lumbar puncture (rare).
- Chronic disease with lower risk of visual loss. May have small fluctuations in disease activity, frequently with weight changes (the majority).
- Those in remission and off treatment

Mollan SP, et al. Pract Neurol 2014;0:1–11

- Recurrence may occur in 8-38% of patients weeks to years ff recovery from initial presentation or a prolonged duration of stability. Weight gain associated with recurrence.
- In children , high papilledema grade on presentation is predictive of poor visual outcomes. ($p<0.001$) Vision loss is associated not only with optic atrophy but also with photoreceptor damage($p<0.0001$)

Sidney M Gospe III et al Br J Ophthalmol. 2016 Apr;100(4):505-9

FACTORS ASSOCIATED WITH WORSE VISUAL OUTCOME

Male gender

Black race

Morbid obesity

Anaemia

Obstructive sleep apnea

Acute onset of symptoms and signs of raised ICP

In children , high papilledema grade on presentation is predictive of poor visual outcomes. ($p<0.001$) Vision loss is associated not only with optic atrophy but also with photoreceptor damage($p<0.0001$)

Sidney M Gospe III et al Br J Ophthalmol. 2016 Apr;100(4):505-9

WHAT'S NEW

- Use of octreotide, a growth hormone and insulin- like growth factor inhibitor being evaluated
- Transverse venous sinus stenosis stenting(TSS) – based on theory that IIH patients may have stenosis of the transverse sinus or other cerebral vein
  - Decreases cerebral venous pressure - increased CSF absorbtion and decreased ICP
  - Side effects stent migration, venous sinus perforation, instent thrombosis, subdural haemorrhage, recurrent stenosis proximal to stent

SUMMARY

- Idiopathic intracranial hypertension is a diagnosis of exclusion where NO cause can be found.
- Characterized by symptoms and signs of raised intracranial pressure and papilloedema in the absence of a space occupying lesion.
- Some drugs and systemic conditions may predispose to IIH
- Evaluation involves a good history, ocular examination to determine presence of papilloedema and rule out any underlying cause.
- Neuro imaging MRI/MRV
- A Lumbar puncture should be performed to rule out any intracranial mass lesion, analysis of CSF for inflammatory or neoplastic causes as well as for the opening pressure
- Treatment is aimed at alleviating symptoms of raised intracranial pressure and preserve vision and involves a multidisplinary approach involving a dietician, neuro- ophthalmologist, neurologist and neurosurgeon.

ACKNOWLEDGEMENT

All who have helped in diverse ways to make this possible but especially

My mentee Vera Beyou

My mentor Vera Essuman

THANK YOU